Communicable Disease BULLETIN

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New Hampshire Surveillance for Creutzfeldt-Jakob Disease

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Case Report

A 63 year-old woman was admitted to a community hospital in Connecticut with a four-week history of progressive confusion and agitation. The patient lived alone and reportedly had some decline in her memory for the past six months. Her ability to function had declined progressively during the last six to ten weeks before hospitalization. On admission the patient's neurological exam was significant for an awake but mute state. She did not respond to any verbal stimuli or follow any commands. Occasional muscle jerks in upper extremities were noted. Evaluation with laboratory testing, cerebrospinal fluid, CT scan and MRI of the brain was unremarkable. She was transferred to a community hospital in New Hampshire eight days after her initial admission for further medical evaluation and treatment. She was diagnosed clinically as progressive dementia without clear etiology. The patient died 22 days after initial hospitalization. A limited brain-only autopsy was performed and her final pathologic diagnosis was: *Primary – Extensive spongiform degeneration consistent with Creutzfeldt-Jakob disease (CJD); Secondary – Changes consistent with senile dementia, Alzheimer's type.*

History and Background

CJD is not a new infectious disease, having been recognized for the better part of a century. Two German physicians, after whom the disease was named, first identified it in the 1920's.[1] The first recognized transmissible spongiform encephalopathies (TSE), sheep scrapie, was reported by sheepherders in the mid-18th century. Mad cow disease which was first recognized in 1986, appears to have originated from scrapie, a naturally occurring disease of sheep and goats. There is strong evidence and general agreement that mad cow disease resulted from the feeding of rendered scrapie-containing sheep meat-and-bone meal to cattle. It has since been recognized in most sheep-breeding countries and is widespread in the United Kingdom where until 1988 the rendered carcasses of livestock (including sheep) were fed to other animals as a protein-rich nutritional supplement. It has not occurred in the United States or other countries that have historically imported little or no live cattle, beef products or livestock nutritional supplements from the United Kingdom.[2] Humans have likely been exposed to scrapie by eating sheep meat, although there is no evidence that CJD in humans has been associated with scrapie.[3]

CJD is one of a group of diseases called TSEs, that also includes kuru, Gerstmann-Straussler-Scheinker disease, fatal familial insomnia, scrapie of sheep, and mad cow

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Table 1. *Types of Creutzfeldt-Jakob disease*

Classic sporadic CJD

- Etiology – occurs spontaneously without identifiable cause
- Age onset – between the ages of 50 and 70
- Early symptoms – minor lapses of memory, confusion, mood changes, loss of interest
- Psychiatric symptoms – most commonly depression or less often a schizophrenia-like psychosis
- Neurological signs – unsteadiness, difficulty walking, involuntary movements, clumsiness, blurred vision, and slow or slurred speech develop as the illness progresses
- Time course – median of 4.5 months after onset of neurological findings until death

Iatrogenic CJD

- Etiology – accidental transmission of classic sporadic CJD as a result of a medical procedure. Sources are contaminated instruments insufficiently autoclaved after use in a patient with CJD
- Signs and symptoms – similar to those in classic sporadic CJD

Familial CJD

- Etiology – occurs genetically in families
- Age onset – slightly younger age than either classic of iatrogenic CJD
- Time course – more protracted course than classic sporadic CJD

New variant CJD

- Etiology – linked to mad cow disease by consumption of infected beef
- Age of onset – mean age at onset is 29 years vs. 60 years for classic CJD
- Early symptoms – anxiety and depression
- Neurological symptoms – uniformly associated with incoordination and a type of sensory impairment of touch, not typical of classic CJD
- Time course – average of 14 months from onset of neurological symptoms to death

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disease of cattle and dairy cows. There are four types of CJD: classic sporadic, iatrogenic, familial and new variant. In general, all four types have long, months to years, incubation periods preceding the onset of clinical illness. The disease usually begins with progressive mental deterioration that soon becomes associated with progressive unsteadiness and clumsiness, visual deterioration, muscle twitching and a variety of other neurological and psychiatric signs and symptoms. The patient is usually mute and immobile in the terminal stages, and in most cases death occurs within a few months of onset of symptoms (Table 1).[4]

The diagnosis of CJD is a challenge. Most routine laboratory or diagnostic studies are of little value. Definitive diagnosis requires biopsy and examination of brain tissue pre or post mortem. No effective treatment is available and there are no known cases of remissions or recoveries.

Transmissible spongiform encephalopathies are caused by the progressive accumulation in the central nervous system of a structurally abnormal form of a normal protein known as the prion protein. This "infectious agent" or prion protein, interacts, and damages normal proteins within brain cells. Gaps in brain tissue develop giving the brain a characteristic "sponge-like" appearance when viewed under a microscope.[5]

Media attention to the recent outbreaks of mad cow disease in the United Kingdom and the association with new variant Creutzfeldt-Jakob disease (nvCJD) has attracted worldwide attention. New variant Creutfeldt-Jakob disease of humans is hypothesized to be caused by consumption of BSE contaminated beef. This type of CJD represents the first recognized instance in which a TSE of animals has crossed a species barrier to infect humans. There have been no documented cases of nvCJD or mad cow disease in North America.

Epidemiology

CJD occurs worldwide at an estimated annual incidence rate of 0.5 to 1.5 cases per million population with no seasonal or geographic predisposition except for areas of high familial occurrence. To date there is no evidence of person-to-person transmission among family members. Although not a reportable disease in most states, the possibility that mad cow disease can be spread to humans has focused increased attention on national CJD surveillance. The Centers of Disease Control and Prevention (CDC) monitors trends and the incidence of CJD in the United States by analyzing death certificate information compiled by the National Center for Health Statistics. These data indicate that there is no discernable increase in the incidence of CJD detected in the United States during the past two decades. The annual CJD death rates in the United States from 1979-1995 have been stable ranging between 0.8 cases per million in 1980 and 1.1 cases per million in 1987.[6]

New Hampshire is one of several states that does not require reporting of patients with CJD. However, New Hampshire does participate with CDC in monitoring death certificates on all cases of CJD. Based on a national rate of 1 case per million persons per year, approximately 1 to 2 cases of CJD are expected in New Hampshire each year. Listed below is a summary of death certificate data from the New Hampshire Vital Records Database.

Table 2. *Numbers of deaths attributed to Creutzfeldt- Jakob disease. New Hampshire, 1993-1999.*

	# of deaths
1993	0
1994	0
1995	2
1996	3
1997	5
1998	3
1999	4
Total	17

The true prevalence of CJD in the United States is unknown. Case recognition and the challenge of diagnosis are contributing factors. In addition, surgical and autopsy procedures to determine etiology of symptoms are rarely performed on patients with atypical dementias or in patients in whom the diagnosis may have been considered because of the fear of transmission of "infection". Performing limited autopsies on the brain only or brain biopsies of suspect cases is the only way to monitor the true prevalence of CJD and the emergence of nvCJD in the United States. Diagnostic testing specific to the identification of CJD is performed at designated laboratories in the United States. The Bureau of Communicable Disease Control will assist in the sending of any brain specimens to the appropriate laboratory for the purpose of identifying CJD.

Stringent recommendations from the World Health Organization for decontaminating instruments or equipment that have come in contact with tissues of patients with CJD during surgical or autopsy procedures are now used uniformly throughout the United Kingdom and other countries. These recommendations, based on proven studies effective in the decontamination of the prion protein, are available in full text on the Internet at http://www.who.int/emc-documents/

For more information on CJD in New Hampshire and transportation of brain specimens for diagnostic testing, please contact the New Hampshire Bureau of Communicable Disease Control at (603) 271-4496 or 1-800-852-3345 extension 4496.

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Diabetes in New Hampshire, 1995-1998

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[Note: This issue of the Communicable Disease Bulletin *is including the following chronic disease topic highlighting epidemiology in this area. We hope to include further topics such as this as we consider changes to the publication.]*

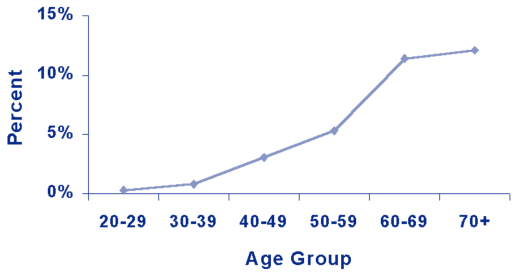
Diabetes is one of the leading causes of disability and death in the United States. It is a leading cause of blindness, end-stage renal disease, lower limb amputation, and impotence.[1] Heart disease and stroke are two to four times more common in persons with diabetes.[1] It is estimated that the national health care costs for diabetes were $98 billion dollars in 1997.[1] In New Hampshire, diabetes is present in 12% of all hospitalizations and is the 6th leading cause of death. To better understand diabetes as a public health problem in New Hampshire, we examined data from the Behavioral Risk Factor Surveillance System (BRFSS).

The BRFSS is a random-digit-dialed telephone survey of the civilian, non-institutionalized population aged ≥18 years. From 1995 through 1998, approximately 1,500 New Hampshire residents participated in the survey each year. Results from four years were aggregated to increase the number of persons with diabetes for this analysis.

During the four-year period, 4.1% of BRFSS respondents in New Hampshire reported being diagnosed with diabetes. This is somewhat lower than the national estimate for 1997 of 4.8%. Diabetes prevalence increased with increasing age from 0.3% among those 20-29 years of age to 12.1% among those ≥70 years old (Figure 1). Based on the BRFSS estimate, there were an estimated 36,600 adults in New Hampshire who had been diagnosed with diabetes. Fifty-nine percent of persons with diabetes in New Hampshire were ≥60 years of age.

When assessing risk factors, 70% of persons with diabetes in New Hampshire reported being overweight (Body Mass Index ≥25.0 (BMI=weight [kilograms]/height [meters^2])) compared to 50% of the rest of the population (Figure 2). Fifty-seven percent of persons with diabetes reported having hypertension compared to 20% of rest of the population. Persons with diabetes also reported a higher prevalence of elevated cholesterol (46%) compared to persons without diabetes (28%). Forty-five percent of persons with diabetes reported no leisure time physical activity during the past month compared to 24% of the rest of the population. Persons with diabetes reported a lower prevalence of smoking than did the general population (16% versus 24%).

Figure 1. *Prevalence of Diabetes by Age Group. New Hampshire, 1995-1998.*



The BRFSS data were also used to measure if persons with diabetes received appropriate preventive clinical services. Seventy-one percent of persons with diabetes reported receiving a dilated eye examination during the past 12 months, 69% had had at least one foot examination during the past 12 months, 47% had been immunized against influenza during the past year, 38% had ever received an immunization for pneumococcal disease, and 19% reported having had at least one glycosylated hemoglobin (e.g., hemoglobin A1C) test during the past 12 months. Ideally, foot exams and glycosylated hemoglobin testing should be done quarterly. Only 32% of persons with diabetes had at least four foot exams during the previous twelve months. For glycosylated hemoglobin, only 10% had been tested at least four times during the previous year.

The results of the BRFSS survey have at least three limitations. First, the sample size for persons with diabetes is too small to look at results for individual years. The data need to be aggregated over several years for meaningful analysis. Second, the prevalence of diabetes is most likely an underestimate as many persons with diabetes remain undiagnosed. Finally, recall bias may affect some of the results. For example, many persons with diabetes are unfamiliar with the term "hemoglobin A1C" and may not know if they were tested by their physician during the preceding year.

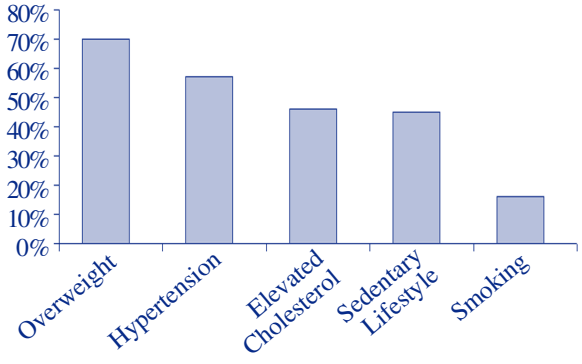
The population of the New Hampshire is projected to increase 17% over the next 15 years. However, the population of persons ≥60 years of age is expected to increase 67% during the same time period due to the aging of the baby boomers' generation and increased life expectancy. As the population ages and the prevalence of overweight and sedentary lifestyle increases, the number of persons with diabetes in New Hampshire is likely to increase substantially.

Attempts to prevent and control diabetes require a comprehensive strategy. Primary prevention consists of maintaining appropriate body weight, good nutrition, and adequate amounts of physical activity. Secondary and tertiary prevention consists of good glucose control and interventions such as immunizations and foot and eye exams. For copies of the *New Hampshire Guidelines for Diabetes Care* or more information on the New Hampshire Diabetes Education Program, please call (603) 271-5173.

References

1. Centers for Disease Control and Prevention. National Diabetes Fact Sheet: National estimates and general information on diabetes in the United States. Revised edition. Atlanta, GA: U.S. Department of Health and Human Services, Centers for Disease Control and Prevention; 1998.

Figure 2. *Prevalence of risk factors among persons with diabetes. New Hampshire, 1995-1998.*



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1. Brown P, Bradley R. 1755 and all that: A historical primer of transmissible spongiform encephalopathy. *BMJ* 1998; 317:1688-92.
2. Brown P, et al. Bovine spongiform encephalopathy and variant Creutzfeldt-Jakob disease: Background, evolution, and current concerns. *Emerging Infectious Diseases* 2001; 7(1).
3. Orriss GD. Animal diseases of public health importance. *Emerging Infectious Diseases* 1997; 3(4): 497-502.
4. Whitley RJ, et al. Technical report: Transmissible spongiform encephalopathies: A review for pediatricians. (T109906). *Pediatrics* 2000; 106(5): 1160-1165.
5. Pruisner SB. Prion diseases in humans and animals. *Virology* 1996; 7: 159-73.
6. Bovine spongiform encephalopathy (BSE) in the United Kingdom and Creutzfeldt-Jakob disease (CJD) in the United States. CDC Website. Available at http://www.cdc.gov/ncidod/diseases/cjd/qa96bse.htm

Communicable Disease Reports – No Fax Policy

Public health disease surveillance systems collect essential epidemiological information on many diseases and conditions. At the public health department, assurance of patient confidentiality is our highest priority. Therefore, we continuously evaluate our system in order to maintain an exceptional level of system confidentiality and security with all our public health disease surveillance data. Our evaluation process serves to enhance our existing practices and procedures.

We do wish to inform New Hampshire health care providers of the Division of Epidemiology and Vital Statistics 'no fax policy' for all HIV/AIDS surveillance data, both incoming and outgoing communication. HIV/AIDS information includes: HIV antibody tests, HIV viral load tests, CD4 lymphocyte counts and *Pneumocystis carini* pneumonia reports. Alternatively, we request that all HIV/AIDS information either be confidentially telephoned or mailed to:

STD/HIV Surveillance Coordinator
Bureau of Communicable Disease Surveillance
6 Hazen Drive
Concord, NH 03301 Tel: (603) 271-3932

Additionally, we restrict all outgoing fax communication for all other communicable disease information containing patient identifiers. With the exception of tuberculosis investigation, outgoing communicable disease information may only be faxed to specific pre-programmed numbers. All (non-HIV) communicable disease information may continue to be reported by fax and will be received by a secure fax machine with restricted access. All confidential communications to out of state health departments are conducted by telephone or by mail.

Our Department appreciates your ongoing support of public health disease surveillance. If you have any questions or concerns about disease reporting please call (800) 852-3345, X 0279 in New Hampshire or (603) 271-0279.